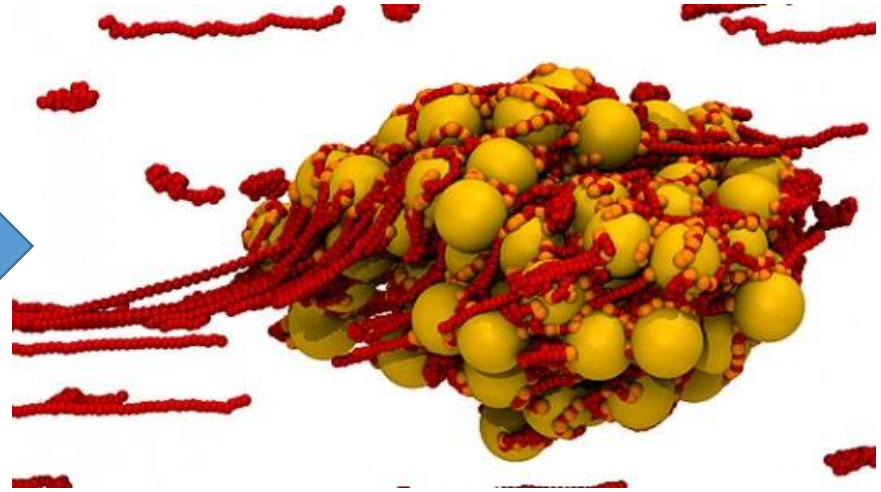
Bleeding and Gastrointestinal Disorders
Victory Junction Medical Staff and Volunteer Training

Bleeding Disorders

Bleeding Disorders

The process of blood clotting involves platelets and several plasma proteins, called coagulation or clotting factors.

Platelets, a type of blood cell, stick together.



Clotting factors work together in a cascade to form a fibrin clot.

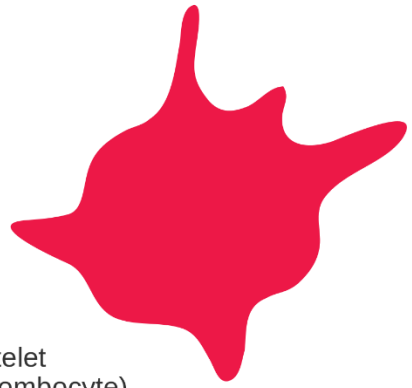
In a person with a bleeding disorder, some part of this process is missing or doesn't work and this can lead to excessive bleeding.

Bleeding Disorders

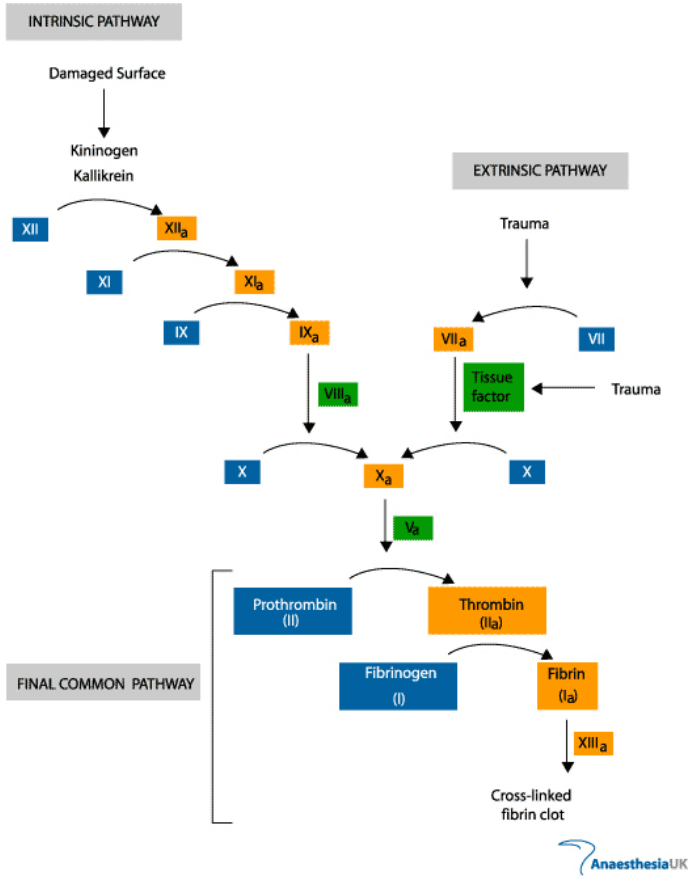
There are many, many different bleeding disorders but most fall into one of two categories.

Platelet Disorders

- Acquired vs. congenital
- Idiopathic thrombocytopenic purpura (ITP)



Platelet (thrombocyte)



Clotting Factor Disorders

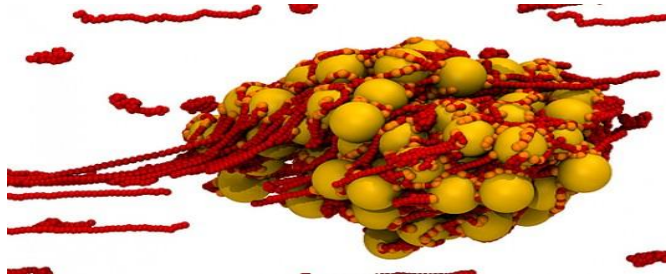
- Hereditary
- Factor deficiencies
- Von Willebrand
- Hemophilia
  - A
  - B

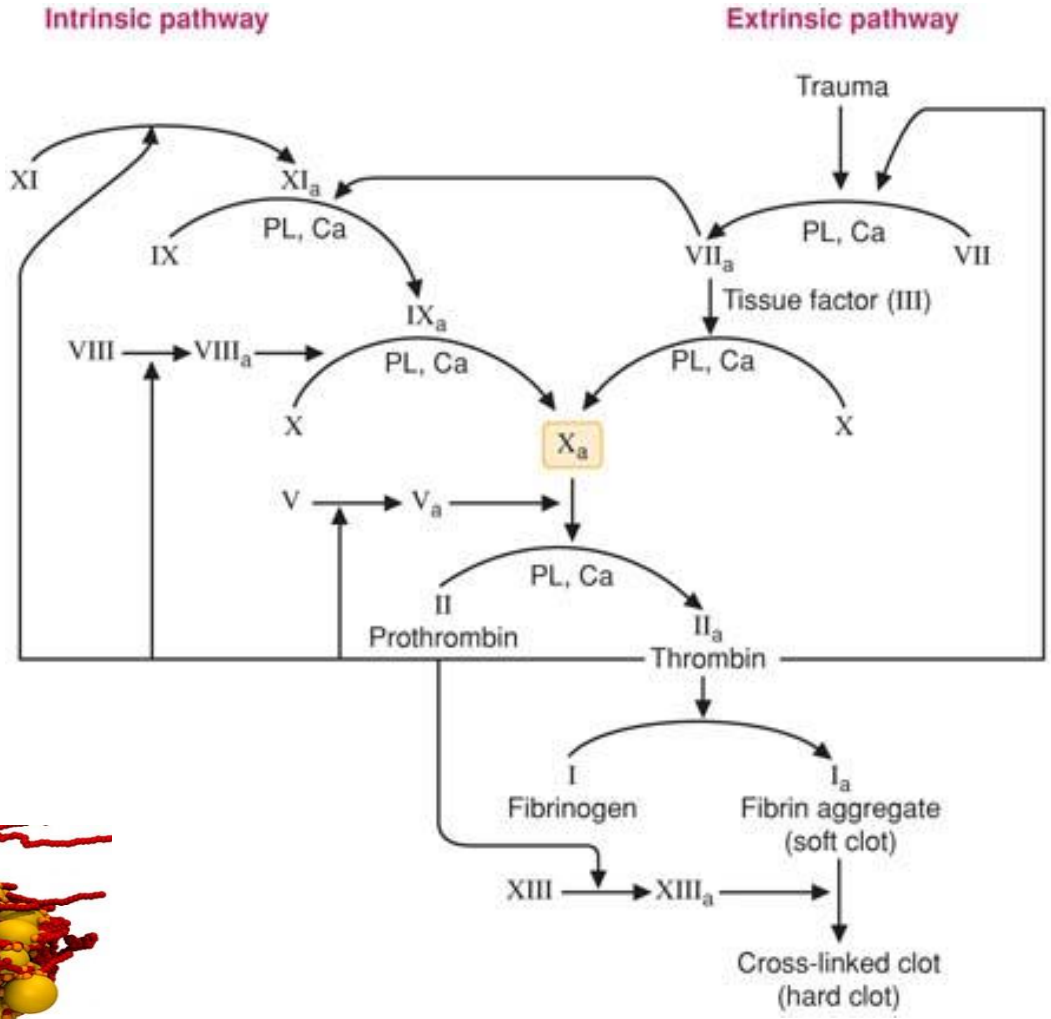
Bleeding Disorders – Clotting Cascade

The clotting cascade is initiated with injury to tissue or blood vessels.

Platelets stick together to form a "plug" at the site.

The cascade, like a chain reaction, requires several clotting factors, tissue factor, thrombin and fibrin to transform the loose platelet plug into a secure, hard clot.

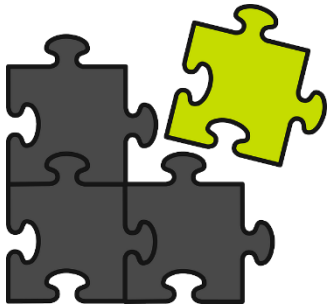




Bleeding Disorders – Different types of bleeds

A bleeding disorder presents differently depending on the component of the clotting cascade that is affected. Think about a puzzle – it looks different depending on which piece is missing.

Bleeding with low ***platelets*** (thrombocytopenia) or platelet dysfunction



- Slow “plug” formation
- Mucosal bleeding
  - **Nosebleeds are common**
  - Gum bleeding
  - Girls with ITP/VWF often have excessive menstrual bleeding

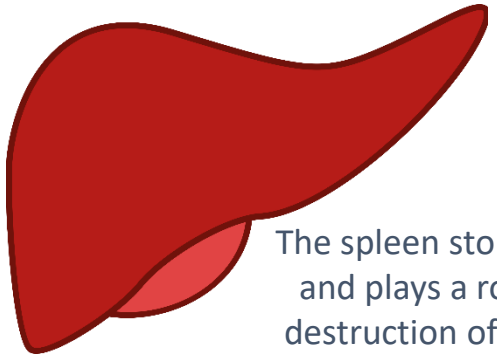
VWD bleeding acts more like platelet bleeding because of how Von Willebrand factor works.

Bleeding with ***factor*** deficiency

- A “plug” forms but doesn’t become a strong clot = “leaky faucet”, slow bleeds
- More concern for **intracranial bleeding**
- Less visible bleeding
- **Muscle and Joint bleeding**
  - Same joint can be affected multiple times
  - Long term damage

Bleeding Disorders – ITP

Idiopathic thrombocytopenic purpura (ITP) is an autoimmune disorder in which platelets are destroyed by autoantibodies binding to platelet antigens.



The spleen stores, filters, and plays a role in the destruction of platelets. Removing the spleen used to be the primary treatmen for ITP, although this is no longer recommended.

In children, ITP may present 4-6 weeks after an acute viral illness.

It is thought that the viral illness plays a role in activating the autoimmune process.

ITP is characterized by persistently **low platelet counts (<30,000),** and typically presents with evidence of bleeding (i.e. petechiae, purpura, nosebleeds, gum bleeding, etc.) ITP can be either acute or chronic.

Normal platelet counts range from 135-145,000. At levels <10,000, there is increased risk for spontaneous hemorrhage.

Bleeding Disorders – ITP

Acute ITP

- Most cases in children are acute and self-limiting
- Approximately 80% spontaneously recover within 6 weeks
- More likely to occur after viral illness
- Usually only treated if child is symptomatic (bleeding)
- Anticipatory guidance for parents is important, as child is often cared for at home not hospital

Chronic ITP

- ITP consider chronic if “relapsed” or refractory 6 months from initial presentation
- More likely if child is >10 y.o. at diagnosis
- More common in adolescent females
- Diagnostics include tests to rule out other causes of thrombocytopenia, including autoimmune diseases, bone marrow failures, cancers
- Treatment similar to acute ITP, but more difficult to manage long-term

ITP Treatment

Supportive care. Bleeding precautions necessary for children with platelet count <20,000.

Treatments target suppressing the overactive immune response in ITP:

- Corticosteroids - least expensive, effective, but have side effects
- IVIG infusions – effective in >80% of children, often first line therapy for symptomatic ITP
- New medications to stimulate production of platelets – expensive, mixed results, side effects limit use
- Platelet transfusion only recommended for intracranial hemorrhage or life threatening bleeding

Guidelines for the investigation and management of idiopathic thrombocytopenic purpura in adults, children and in pregnancy. (2003). British Journal of Haematology

Bleeding Disorders – Factor Deficiencies

Hemophilia is an inherited disease in which a person's blood doesn't form clots properly because of low levels of a clotting factor.

Each clotting factor is a protein that plays an important role in the clotting cascade.

Hemophilia A is about 4 times as common as hemophilia B. VWD is NOT hemophilia, but is a common factor deficiency bleeding disorder.

Hemophilia A

"Classic Hemophilia"

Factor VIII deficiency

Hemophilia B

"Christmas disease"

Factor IX deficiency

Von Willebrand Disease

Types 1, 2, 3

Von Willebrand factor deficiency

Hemophilia is classified as mild, moderate, or severe depending on the degree of deficiency.

http://www.cdc.gov/ncbddd/hemophilia/facts.html

Bleeding Disorders – Von Willebrand Disease

Platelet
(thrombocyte)

Von Willebrand factor attaches to platelets and helps them stick together to form the initial "plug."

VWD is the most common bleeding disorder in the U.S., affecting about 1% of the population.

Type 1 VWD

- Most common type, least severe
- Low levels of VWF
- May also have low levels of factor VIII

Type 2 VWD

- Normal level of VWF, but it doesn't work
- Various subtypes

Type 3 VWD

- Most severe type
- Minimal or no VWF
- Low levels of factor VIII

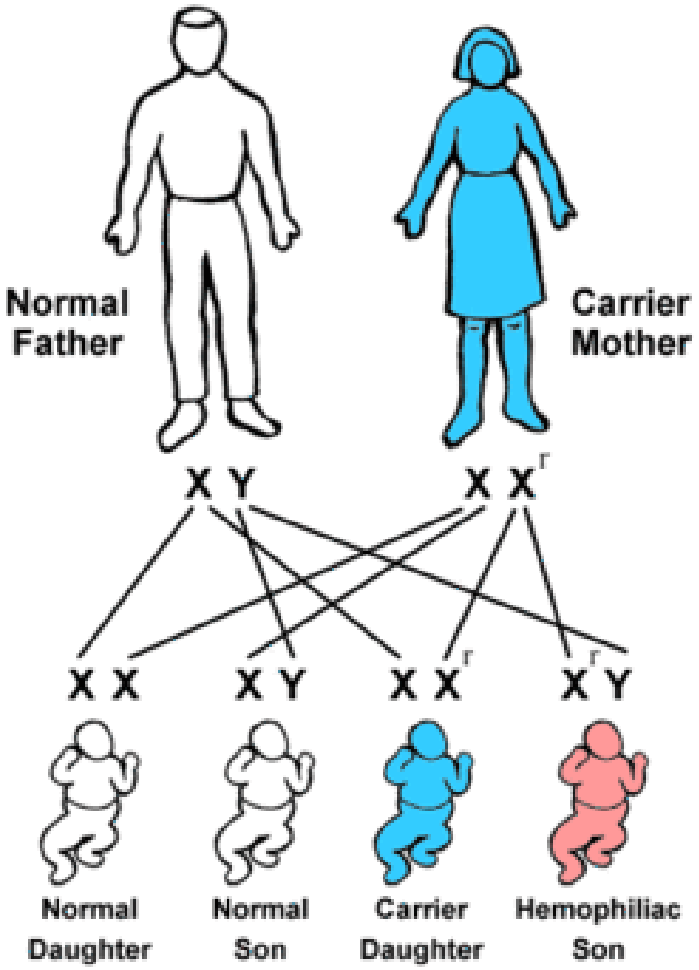
Von Willebrand disease occurs in equal rates in males and females, and is usually inherited.

Nosebleeds are a common symptom, as is heavy menstrual bleeding in females.

Treatments include DDAVP, oral contraceptives (for women), and other treatments similar to hemophilia.

http://www.cdc.gov/ncbddd/vwd/facts.html

Bleeding Disorders – Hemophilia - Demographics



Hemophilia is inherited by a gene on the X chromosome.

Females are carriers of the gene, but males are affected by the disease if the “X” inherited has the gene.

According to the CDC, about 400 babies are born with hemophilia each year in the U.S., or 1 in 5,000 male births. It is diagnosed by a special blood test.

A female can have hemophilia, but it is very rare.

Hemophilia affects people from all racial and ethnic groups.

http://www.cdc.gov/ncbddd/hemophilia/data.html

Bleeding Disorders – Hemophilia - Treatment

In all types of hemophilia, the treatment is similar – ***replace the missing factor.***

Factor is administered as an IV infusion, usually over a few minutes.

People with hemophilia often learn to self-infuse their factor.

For severe hemophilia, prophylactic factor doses maintain adequate levels to prevent spontaneous bleeds.

If minor or major injury occurs, additional factor is given to stop bleeding.

Factor is derived from human plasma or genetically engineered.

All factor products are carefully screened for viruses.

http://www.cdc.gov/ncbddd/hemophilia/treatment.html

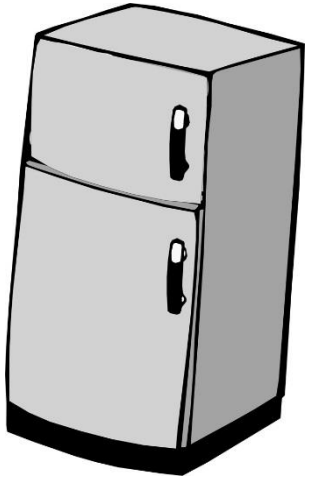
Bleeding Disorders – Hemophilia - Treatment

Factor is dosed in international units (IU) and in a range.

- Because factor is very expensive, it is important not to waste.

- Factor comes as a powder and solution for reconstitution immediately before administration.

- People with hemophilia often use a “dose range” but will round the actual dose to use the entire vial.



- Factor often needs to be stored in a refrigerator or cooler with ice packs.

http://www.cdc.gov/ncbddd/hemophilia/treatment.html

Bleeding Disorders – Hemophilia -Treatment

Typically, unit nurses are responsible for factor administration. As a volunteer, you are welcome to assist if you are comfortable or want to learn about factor.
Specialists providers, full time staff, and the campers themselves have a wealth of knowledge!

Prophylactic factor infusions will take place in the Body Shop before breakfast. Additional PRN infusions may be needed throughout the week.

Administering this medication at camp:

- Verify the 5 rights of med administration!
- Reconstitute the medication (allow/encourage the camper to help with supervision)
- Draw up the medication into a syringe once fully dissolved (but don't shake)
- Infuse with a butterfly needle (The camper may participate by tying/untying the tourniquet, pushing the syringe, and with the stick. Some campers will self-infuse or want to learn to self-infuse with supervision.)
- Dispose of butterfly in sharps container. The camper's family may track lot numbers from the vial.

Helpful Hint: The factor brand name usually tells which factor it replaces.
NovoSeven – factor VII
Advate, Kogenate, Recombinate, Helixate – factor VIII ("ate")
BeneFix – factor IX

Bleeding Disorders – Hemophilia -Treatment

Occasionally, people with a bleeding disorder will take other medicines to prevent or control bleeding. These can be used daily or PRN for bleeding.

Aminocaproic acid ("Amicar") prevents an existing clot from breaking apart. This is usually used for mucosal/oral bleeding, and can be given as a tablet, liquid, or "swish and spit" suspension.

Desmopressin acetate ("DDAVP") causes factor VIII to be released from body tissues where it is stored. It is more often used in treatment for Von Willebrand disease or mild hemophilia. This is usually given as a nasal spray or pill.

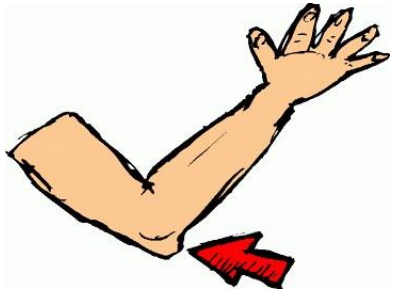
http://www.cdc.gov/ncbddd/hemophilia/treatment.html

Bleeding Disorders – Hemophilia - Inhibitors

An ***inhibitor*** is an antibody that person's body creates against the clotting factor.

The inhibitor prevents the factor from working, which can lead to more bleeding problems.

This happens in about 15-20% of people with hemophilia.



- Hemophiliacs with inhibitors are more likely to have joint problems related to bleeding.
- Treating hemophilia with inhibitors can be very expensive and very difficult.

http://www.cdc.gov/ncbddd/hemophilia/facts.html

Bleeding Disorders at Camp

No contact sports

- Modify activities to avoid potential joint injury or head injury.

Forbidden OTCs

- **NSAIDs and aspirin are contraindicated with a bleeding disorder.**

Medical staff should be notified **immediately** of any injury in a camper with a bleeding disorder.

Prophylactic Treatment

- Camper's with hemophilia are often prescribed a prophylactic dose of factor before high-risk activities, like the adventure tower.
- Make sure counselors know to bring these campers to the Body Shop in the morning.

Nosebleeds

- Nosebleeds are a common concern for campers with bleeding disorders.
- Apply pressure, ice, and if the nosebleed lasts > 20 minutes, notify medical staff.
- Because clots form more slowly, they can be large. Do NOT have the camper blow their nose or dislodge the clot.

Bleeding Disorders – Emergencies/Injuries at Camp

- **Head injury** in a camper with hemophilia requires immediate attention. Perform a quick neuro check, and **notify the provider on call.**

- **Joint injury:** The camper may have a "target joint" where he tends to have bleeds. Injury, pain, swelling, or stiffness in any joint should be assessed by a provider.
  - NO NSAIDs, NO ASPIRIN!
  - Ice packs are fine. R.I.C.E. is advised in addition to dosing factor per orders.

If you think your camper may have a bleed, **notify the provider on call**. Each brand of factor and type of injury calls for a different calculation for "factor correction."

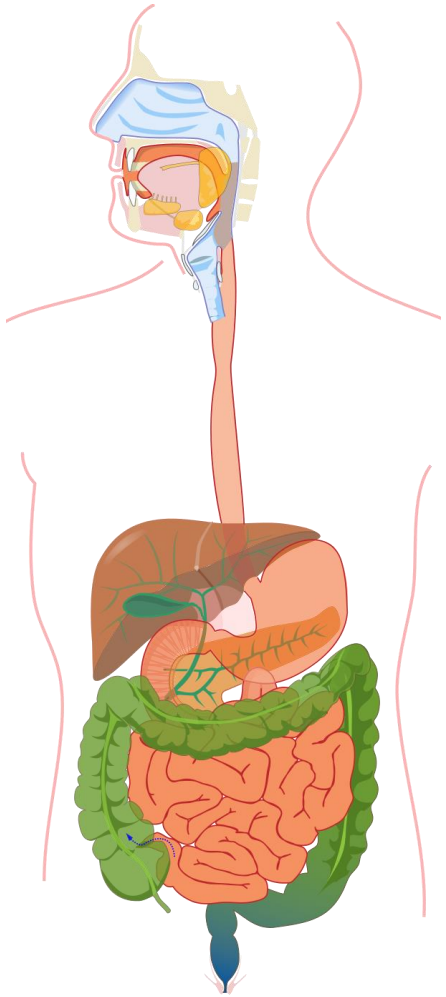
At medical check-in, ask your camper:
Do you have a target joint?
Have you had any injuries recently?
Do you have any swollen or painful spots today?
Who usually does your factor infusion?

Things to Remember…

- Every child is unique. Do your best to get to know your campers and listen to them!
- We do our best to stick to campers' home routines. Medications and treatments should be done the way they are done at home as much as possible.
- As a volunteer, you are not expected to be an expert in everything. We welcome your knowledge and experience and aim to work as a team!
- The unit nurses, full time staff, and specialist providers are excellent resources and here to help. Always ask if you are unsure or concerned about a medication, treatment, or camper's condition.

GI Disorders

GI Disorders



The gastrointestinal system, including the GI tract, liver, gallbladder, and pancreas, digests the food we eat into useful energy and waste products with a wide variety of enzymes, bacteria, and regulating nerves and hormones.

Campers may have a variety of GI disorders, including:

- Celiac disease
- Crohn's disease
- Ulcerative Colitis
- Inflammatory Bowel Disease (IBD)
- Gastroparesis (or other functional GI disorders)
- Hirschsprung's
- Liver or small bowel transplant
- Congenital Sucrase Isomaltase Deficiency (CSID)
- Eosinophilic esophagitis (EOE)

GI Disorders – Celiac Disease

- Celiac disease, which occurs in about 1% of the population, is an autoimmune disease in which the person's body cannot tolerate gluten.

- Consuming the protein gluten damages intestinal villi in people with Celiac disease, causing intestinal inflammation and decreased ability to absorb nutrients.

Symptoms and complications of Celiac disease:

- Bloating
- Diarrhea/constipation
- Stomach pain
- Nausea and vomiting
- Weight loss, or failure to thrive in children
- Irritability

The treatment for Celiac disease is a gluten-free diet. At Victory Junction, the NUTrition team will prepare foods that are safe for these campers.

Celiac disease occurs in higher rates among those with other autoimmune diseases, such as type 1 diabetes, rheumatoid arthritis, or Addison's disease.

National Institute of Diabetes and Digestive and Kidney Diseases. (2015). Celiac disease. Retrieved from http://www.niddk.nih.gov/health-information/health-topics/digestive-diseases/celiac-disease/Pages/facts.aspx

GI Disorders – Crohn's/Ulcerative Colitis/IBD

	Crohn's Disease	Ulcerative Colitis	Inflammatory Bowel Disease
	Chronic disease causing inflammation of the entire GI tract, possibly autoimmune nature	Chronic disease causing inflammation and ulceration of the lining of the large intestine (colon), possibly autoimmune component	Includes both Crohn's and Ulcerative colitis
Common signs and symptoms	Diarrhea, abdominal cramping and pain, weight loss	Diarrhea with blood or pus, abdominal discomfort	Severe diarrhea, constipation, or both
Common treatments	No cure. Medications (immunomodulators, steroids, aminosalicylates, biologic therapies) to reduce inflammation and immune response. Surgery to remove sections of the small or large bowel to relieve symptoms. Good nutrition, low fiber diet.	Medications (immunomodulators, steroids, aminosalicylates, biologic therapies) to reduce inflammation and immune response. Surgery to remove the entire colon can "cure" UC. Good nutrition, low fiber diet.	Balanced diet, medications to reduce inflammation or suppress the overactive immune system. In some cases, surgery.
Complications	Bowel obstruction, fistulas, anal fissures, ulcers, malnutrition. Increased risk of colon cancer.	Rectal bleeding, dehydration, malabsorption, bone changes, systemic inflammation. Increased risk of colon cancer.	Includes those of Crohn's and UC, can lead to malnutrition due to malabsorption of nutrients.

Caring for campers with GI Disorders

Things to remember when caring for campers with GI Disorders:

- Be mindful of food allergies or restrictions, and that each camper may eat differently. (i.e. popcorn during Nascarnival may be restricted or even not offered, gluten free substitutes will be prepared for campers)
- Food can be a sensitive topic, encourage a healthy diet without pushing campers to eat more or less.
- Make sure campers are having bowel movements! Constipation can happen at camp.
- Make sure campers are hydrated! Diarrhea can lead to dehydration.
- No activity restrictions for campers with feeding tubes, but make sure staff are aware.

GI Disorders

Helpful medical check-in questions for campers with GI disorders:

- Do you have a feeding tube? (Assess site.) Tell me about how you do your feeds.
- Do you have a bowel routine? Tell me about how I can help.
- Tell me how you take your medications. (Full or empty stomach, timing, etc.)
- Have you had any flare-ups recently? How does that look for you? How do you treat it?

Keep in mind that a lot of this information is entered prior to check-in, and simply verify that our records reflect the campers current status.

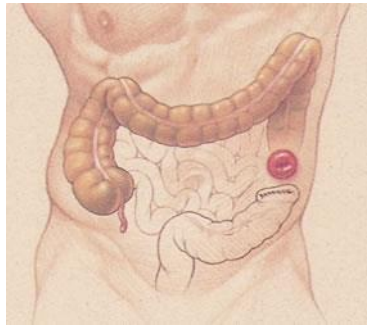
Update the “medical check in” and “unit nurse” notes in CampSite as applicable.

GI Disorders

Some campers may require intervention to have a bowel movement.

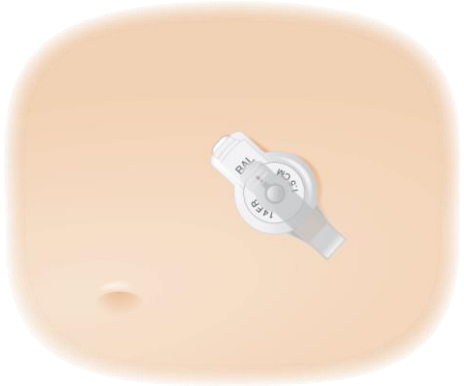
Talk with campers at medical check-in about the importance of having regular bowel movements at camp! It's healthy!

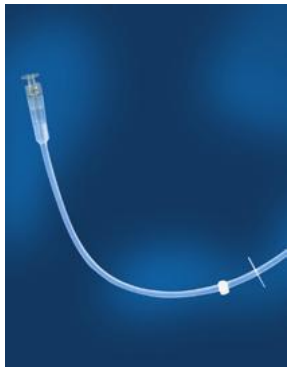
Some campers may have an ostomy to drain stool.

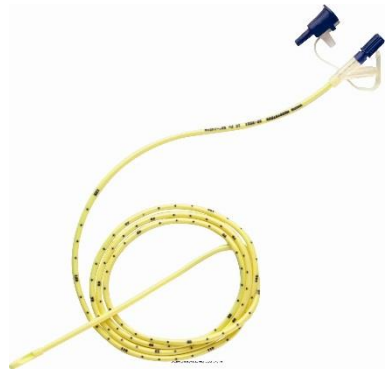


There are no activity restrictions for campers with an ostomy, but a nurse should check it before the camper participates in the waterpark.

Feeding tube care at camp







- Campers may have a feeding tube, such as a G-tube, J-tube, NG, or G-J tube.
- There are no activity restrictions for campers with feeding tubes, but staff should be aware of the tube and avoid any trauma to the insertion site.
- At medical check-in, assess the camper's feeding tube site for any signs of infection or dysfunction, and clarify the home routine for site care and feeding schedule.

Chemotherapy/Biotherapy Precautions

Biotherapy and chemotherapy medications may be prescribed for campers with JIA, cancer, or Crohn's. Some of the most common oral chemotherapy medications for these children are:

- Methotrexate
- Mercaptopurine (6MP)
- Etoposide (Vepesid)

- Methotrexate may also be given subcutaneously

When handling or administering hazardous medications:

- Always wear PPE (gloves)
- Avoid crushing pills (if necessary, notify full time staff or unit nurse)
- Dispose of syringes, etc. in yellow bin
- In case of spills: Contain the area of the spill, alert full time medical staff for cleaning.

Things to Remember…

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- We do our best to stick to campers' home routines. Medications and treatments should be done the way they are done at home as much as possible.
- As a volunteer, you are not expected to be an expert in everything. We welcome your knowledge and experience and aim to work as a team!
- The unit nurses, full time staff, and specialist providers are excellent resources and here to help. Always ask if you are unsure or concerned about a medication, treatment, or camper's condition.

Thank you!

References

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